

MITRAL VALVE PROLAPSE IN BLACKS

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To determine the prevalence and characteristics of mitral valve prolapse in blacks, a population-based sample of 100 black residents of Framingham, Massachusetts, and a series of 15 black patients who had mitral prolapse diagnosed at a tertiary care referral center in New York were studied by uniform echocardiographic and clinical methods. It was found (1) that evidence of mitral valve prolapse occurs in 17 percent of blacks, which is comparable to the highest prevalence estimates in whites; (2) that, as with whites, blacks with evidence of mitral prolapse tended to be young women; and (3) that the high prevalence and variable spectrum of mitral prolapse punctuate the importance of identifying the presumably small but significant subgroup(s) susceptible to the condition's known complications.

Since the original description of mitral valve prolapse by Barlow et al¹ nearly two decades ago, numerous descriptions of this syndrome have been published.²⁻⁶ These have established that mitral valve prolapse occurs in four to seven percent of many different populations.⁶⁻¹² A familial pattern

of occurrence suggesting autosomal dominant inheritance is usually present.¹³⁻¹⁹ Some patients have the clinically recognized mitral valve prolapse syndrome with varied symptoms,²⁰⁻²² whereas an even larger number are found to have asymptomatic mitral valve prolapse.^{22,23} Despite the possibility that major differences might exist between ethnic groups in an inherited condition, the published clinical studies and population surveys have provided little information on the prevalence and characteristics of either symptomatic or asymptomatic mitral valve prolapse in blacks.²⁴

The present study was undertaken to answer two questions: First, what are the frequency and manifestations of mitral valve prolapse in a population-based sample of blacks? And second, do black patients with mitral valve prolapse present with the same spectrum of symptoms and findings as reported in whites?

METHODS

To address the above questions, we have used uniform clinical and echocardiographic methods to assess the prevalence and characteristics of mitral valve prolapse in two populations: a representative sample of the adult black population of Framingham, Massachusetts, and a series of patients with mitral valve prolapse referred to the New York Hospital-Cornell Medical Center in New York City. All subjects were studied after informed consent was obtained.

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Study Subjects

To estimate the prevalence of mitral valve prolapse in blacks, an age-stratified sample of 100 black residents (in the census tract) of Framingham, Massachusetts was studied by history, physical examination, chest x-ray, standard 12-lead electrocardiography, 24-hour ambulatory electrocardiography, and M-mode and two-dimensional echocardiography. To avoid sample bias, a comprehensive list of black Framingham residents was prepared using membership lists of the three predominantly black churches, the National Association for the Advancement of Colored People, men's and women's social clubs, and lists drawn up by blacks active in the community. Each of the 244 individuals identified between the ages of 20 and 70 was assigned a number. Using a random number table, 10 men and 10 women were randomly selected in each of the five age decades (except in the age group of men 60 to 69 years of age, where only nine subjects were identified) and invited to participate.

Sixty-nine of the initial 99 subjects (70 percent) selected came in for evaluation. Thirty-one additional subjects from the total of 244 blacks identified in Framingham were subsequently added, including eleven spouses of invited subjects and 20 individuals selected (in order of previous randomly assigned rank) from the total population. The latter individuals were chosen from age/sex decades which had fewer than 10 subjects scheduled for evaluation. The 100 subjects who came in for evaluation included 55 women whose mean age was 42 ± 12 years (standard deviation) and 45 men of mean age 41 ± 12 years. Table 1 shows the number of subjects in each decade who were identified, who were invited to participate, and who were evaluated.

The second group of 17 blacks studied at the New York Hospital-Cornell Medical Center included 15 identified in a referral population of 137 patients with clinically recognized mitral valve prolapse, and two relatives with mitral prolapse from two separate families. The patients included 13 women of mean age 45 ± 16 years (range 21 to 68 years) and two men aged 29 and 57 years respectively. The family members with mitral valve prolapse were a 28-year-old man and an 11-year-old female.

Patients and family members underwent com-

prehensive evaluation, including a history, physical examination, electrocardiogram, one-dimensional echocardiogram (and two-dimensional echocardiogram in three instances), psychological profile, and recording of dermatoglyphic findings.

Auscultation

All subjects were examined by one or more of the investigators in the supine and left decubitus positions. Auscultation was also performed in the sitting and standing positions. If a midsystolic click with or without a late systolic murmur was not heard with the patient supine or decubitus, handgrip isometric exercise was also employed. The timing and intensity of clicks and murmurs during maneuvers was noted.^{6,25}

Echocardiography

Patients were studied using standard techniques in the partial left decubitus position with the head of the bed elevated 30 degrees. Recordings were made from the third or fourth left intercostal space. Special attention was given to assure that the transducer was perpendicular to the chest wall during imaging of the mitral valve, in order to minimize false positive and false negative diagnoses.^{7,17} Tracings were recorded on light-sensitive paper at 25 or 50 mm/sec. Coded echocardiograms were read independently by two investigators (DDS and RBD). Mitral valve prolapse was diagnosed only when both readers agreed that standard criteria for late systolic or holosystolic prolapse were fulfilled.⁷

RESULTS

Prevalence of Mitral Valve Prolapse in Blacks

Seventeen of the 100 Framingham blacks had typical auscultatory findings (midsystolic click with or without a late systolic murmur) and/or definite echocardiographic evidence of mitral valve prolapse. This total included 13 of 55 women (23 percent) and four of 45 men (9 percent). Fifteen

TABLE 1. DISTRIBUTION OF BLACK SUBJECTS AGE 20 TO 70 YEARS IDENTIFIED AS RESIDENTS OF FRAMINGHAM

	Age (yr)					
	20-29	30-39	40-49	50-59	60-69	Total
Men						
Number of subjects identified	15	38	32	15	9 *	109
Number of subjects invited to participate**	13 (87)	15 (39)	15 (47)	12 (80)	7 (78)	62 (57)
Number of subjects evaluated**	8 (53)	12 (32)	12 (38)	9 (60)	4 (44)	45 (41)
Number of subjects in the initial 10 randomized who were evaluated	6	9	7	8	4 *	34
Number of subjects with mitral valve prolapse (% of those evaluated)	0	1 (8)	2 (17)	0	1 (25)	4 (9)
Women						
Number of subjects identified	27	51	35	11	11 *	135
Number of subjects invited to participate**	14 (52)	15 (29)	22 (63)	9 (82)	9 (82)	69 (51)
Number of subjects evaluated**	10 (37)	12 (24)	18 (51)	8 (73)	7 (64)	55 (41)
Number of subjects in the initial 10 randomized who were evaluated	7	6	8	7	7	35
Number of subjects with mitral valve prolapse (% of those evaluated)	4 (40)	2 (17)	6 (33)	0	1 (14)	13 (24)

*One black man and two black women aged 60-69 were scheduled to come in as part of the Framingham study and their data were not yet available. Thus, only nine black men were randomized

**Percent of total number of subjects evaluated is indicated in parentheses

of 72 subjects (21 percent) aged 20 to 49 years and two of 28 subjects (7 percent) aged 50 to 69 years manifested mitral valve prolapse. A predominance of young women were found in the Framingham blacks with mitral valve prolapse since 71 percent (12/17) of the blacks with mitral valve prolapse came from the 40 percent (40/100) of subjects who were women from 20 to 49 years of age.

Clinical Data

Three of the seventeen (18 percent) Framingham blacks with mitral valve prolapse were symptomatic. All three had nonexertional chest pain, and two of the three also had exertional dyspnea. None of the Framingham blacks with mitral valve prolapse had ST-T or T wave abnormalities in leads II, III or aVF. Only two of 17 Framingham

blacks had any abnormality on the 12-lead electrocardiogram. One, a 43-year-old man, had T wave flattening in the left precordial leads and the other, a 35-year-old woman, had tall T waves (> 10 mm) in the left precordial leads. Two of the 100 Framingham subjects, both with evidence of mitral valve prolapse, had brief, self-limited episodes of ventricular tachycardia during 24-hour ambulatory ECG monitoring. One, a 20-year-old woman, had a midsystolic click followed by a murmur while the other, a 41-year-old woman, had a midsystolic click alone; neither had echocardiographic findings of mitral valve prolapse.

Eleven of the 15 (73 percent) New York hospital patients with mitral valve prolapse were symptomatic: six (40 percent) had nonanginal chest pain; one, a 55-year-old woman, had typical angina; and two had probable angina pectoris; seven had symptomatic episodes of tachycardia, five had ex-

ertional dyspnea; and four had panic attacks while three had possible panic attacks. Four of the referred patients (27 percent) and both family members were asymptomatic. Five of the patients (33 percent) had ST/T abnormalities, but in none were they limited to the inferior leads. One patient, a 30-year-old woman with symptomatic supraventricular tachycardia, had a short PR interval, while a 29-year-old man had a QRS axis of $+105^\circ$.

Auscultatory Findings

Twenty-eight of the 100 Framingham blacks had systolic murmurs. In six of these 28 (21 percent) the murmur was late systolic in timing. Eleven of the 100 Framingham blacks had one or more midsystolic clicks, associated with a late systolic murmur in seven (Table 2).

Eleven of the fifteen New York Hospital referred patients (73 percent) had one or more midsystolic clicks, associated with a late systolic murmur in six. Four had a late systolic murmur alone (27 percent). None of the patients had "silent" prolapse, in contrast to the complete absence of auscultatory findings in one affected family member and the occurrence of only a brief midsystolic murmur at the base in the other.

Echocardiography

Ninety-nine of the 100 Framingham blacks had echocardiograms of sufficient quality to evaluate the mitral valve. Ten of the 99 acceptable echocardiograms showed unequivocal late systolic prolapse (nine patients) or holosystolic prolapse (one patient). Four of the 10 Framingham blacks with echocardiographic mitral valve prolapse had "silent" prolapse with neither click nor murmur (Table 2). Two of the 10 Framingham blacks with echocardiographic mitral valve prolapse had midsystolic clicks and late systolic murmurs; four had midsystolic clicks alone. All of the 15 New York Hospital black patients had echocardiographic prolapse (late systolic in 13); two with borderline echocardiographic findings had midsystolic clicks. One family member had late systolic and one had holosystolic prolapse.

TABLE 2. SUMMARY OF AUSCULTATORY AND ECHOCARDIOGRAPHIC FINDINGS IN 100 BLACKS FROM THE GENERAL FRAMINGHAM POPULATION AND 15 BLACKS REFERRED TO NEW YORK HOSPITAL FOR SUSPECTED MITRAL VALVE PROLAPSE

	Echocardiogram	
	Normal	Prolapse
100 Framingham blacks (Population-based sample)		
Auscultation		
Normal	64	4
Click	2	2
Systolic murmur	19	2
Click-murmur*	4	2
15 New York hospital blacks (Referral population)		
Auscultation		
Normal	0	0
Click	0	5
Systolic murmur	0	4
Click-murmur	0	6

*Excluded from this category is one Framingham subject with a click and murmur whose echocardiogram was technically unsatisfactory

Dermatoglyphics

Six of the 15 New York Hospital patients and one of the two family members had arch patterns in the fingerprints. Overall, 29 of 170 fingerprint patterns (17.1 percent) showed arches.

DISCUSSION

This study shows an unanticipated high prevalence of mitral valve prolapse in a randomly selected subgroup of black women (24 percent) and black men (9 percent) residing in Framingham. Such high prevalences raise the possibility that Framingham blacks may be unrepresentative. A significant effect of selection bias on prevalence in this study is unlikely, however, because the selection was based on randomly assigned numbers. Furthermore, the prevalence of mitral valve prolapse was similarly high in the 69 blacks who were among the initial 99 Framingham blacks selected randomly (six of 35 [17 percent] females

and three of 34 [9 percent] males). This high prevalence is also comparable to the 17.9 percent prevalence of mitral valve prolapse in black South African school children reported by Cohen et al²⁴ but in contrast to the prevalence of 1.4 percent in an earlier report from the same group.²⁶

The prevalence of mitral valve prolapse in blacks in this study is comparable to the highest estimates reported for white women (17 percent, by Markiewicz et al⁷) and for white men (7 percent, by Darsee et al⁸). Most studies suggest that the true incidence of mitral valve prolapse in the white population is lower—4 to 7 percent in males and 6 to 17 percent in women.⁹⁻¹² Our own preliminary findings in similar age whites in Framingham (4 percent in men and 13 percent in women by echocardiography) are in accord with these widely accepted estimates of prevalence. The present findings raise the possibility, requiring evaluation in other populations, that mitral valve prolapse may be even more common among black than white adults. For confirmation this will require further evaluations of other populations.

Comparison of the clinical findings in our two groups of blacks with mitral valve prolapse reveals a dichotomy which has been found in studies of whites. Patients seen in a referral center are predominantly symptomatic, commonly have electrocardiographic abnormalities, with a high concordance between classic auscultatory and echocardiographic findings of mitral valve prolapse. In contrast, individuals discovered to have mitral valve prolapse in the course of a population survey are usually asymptomatic, have few electrocardiographic abnormalities, and commonly present only auscultatory or echocardiographic evidence of mitral valve prolapse. While this discrepancy has led Barlow and others to suggest that different entities are being detected,^{20,21} our finding of totally asymptomatic first-degree relatives with mitral valve prolapse in the families of two of our most symptomatic patients (palpitations, chest pain, dyspnea, stroke, and panic attacks in one; palpitations, chest pain, dyspnea and panic attacks in the other) points to a different possibility; ie, a genetic syndrome with variable expression. These discrepancies and similar variability with regard to symptomatic status among individuals with mitral valve prolapse in white families¹⁸ indicate that mitral valve prolapse has great variability of symptomatic expression even when it is inherited as an

autosomal dominant characteristic within a single family. Patients seen in referral centers and by consultant cardiologists may be selected, by themselves and their physicians to reflect the most symptomatic end of the spectrum of expression, whereas unselected subjects with mitral prolapse may have only a small increase in symptoms.^{23,27} Nevertheless, in subjects with mitral valve prolapse in our population sample, a normal 12-lead electrocardiogram or the lack of symptoms did not eliminate the risk of high grade ventricular arrhythmia. Both of the blacks with ventricular tachycardia on 24-hour ECG monitoring had normal 12-lead electrocardiograms, and one was asymptomatic.

Our findings of the arch pattern in 17 percent of the fingerprints in the New York Hospital blacks with mitral prolapse is comparable to that reported by Schwartz et al in white patients with mitral valve prolapse.²⁸ Control data are lacking on the prevalence of the arch pattern in blacks without mitral prolapse and further studies seem warranted in both blacks and whites to assess whether this pattern is truly associated with mitral prolapse as reported.²⁸

Our finding of a higher prevalence of detectable mitral valve prolapse in young black women is consistent with the findings in whites with regard to both sex and age. Not only has there been a predominance of women in most clinical series^{2,6,14,21} but this has also been reported among first-degree relatives with mitral valve prolapse in family studies.^{15,18} Since the findings in most family studies, as well as the two families evaluated in the present series, indicate that mitral valve prolapse appears to be inherited as an autosomal dominant condition,¹⁴⁻¹⁹ expression of the condition appears to be reduced among both black and white men. Similar to the findings of Fortuin et al,¹⁵ we found a decrease in the prevalence of mitral valve prolapse after age 49 years. While this drop-out could reflect premature mortality, it is more likely, considering the generally benign nature of the condition,^{22,29,30} that the mitral leaflet thickening and scarring which occurs gradually as a secondary change in patients with mitral valve prolapse³¹ causes masking of typical auscultatory and echocardiographic findings.

The high prevalence and variable spectrum of clinical findings of mitral valve prolapse in blacks as well as whites punctuate the importance of

identifying the presumably small but significant subgroups susceptible to the condition's known complications: bacterial endocarditis, progressive mitral regurgitation, rupture of chordae tendineae, sudden death, and cerebral ischemic events. Prospective evaluation of the broad spectrum of subjects with mitral valve prolapse may identify useful clinical predictors of risk. In addition, recent reports of biochemical³² and anatomic³³ heterogeneity in mitral valve prolapse suggest that this syndrome may encompass several phenotypically similar conditions which might differ substantially in risk.³⁴

Acknowledgments

Supported in part by a grant from Smith Kline & French, Inc, to the Association of Black Cardiologists, by grant HL-22006 from the National Heart, Lung and Blood Institute, and by Teacher-Scientist Awards to Drs. Devereux and Brown from the Andrew W. Mellon Foundation.

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